

COMPLETE NEUROLOGICAL RECOVERY AFTER SURGERY FOR MESENCEPHALIC CAVERNOMA: CASE REPORT

Krešimir Rotim, Tomislav Sajko, Ivan Škoro, Marina Zmajević-Schönwald and Marta Borić

Clinical Department of Neurosurgery, Sestre milosrdnice University Hospital Center, Zagreb, Croatia

SUMMARY – Cavernous malformations are classified as a group of vascular malformations of the central nervous system. Conservative treatment of brainstem cavernomas is accompanied with poor outcome. Surgery of brainstem cavernomas still poses a challenge due to the high risk of neurological damage and respectable morbidity. We report a case of complete neurological recovery in a 24-year-old female patient with mesencephalic cavernoma treated surgically. This case highlights that careful microsurgical treatment with the goal of complete cavernoma excision remains the treatment of choice in cases with *de novo* or recurrent hemorrhage. Intraoperative neurophysiologic monitoring should be used as the gold standard during brainstem cavernoma operations in order to avoid nuclear and long tract damages.

Key words: *Hemangioma, cavernous, central nervous system – surgery; Mesencephalon; Hemorrhage; Recovery of function; Case reports*

Introduction

Surgery of brainstem cavernomas poses a challenge due to the high risk of neurological damage and respectable morbidity. Conservative treatment and stereotactic radiosurgery represent the alternative treatment methods but with debatable results. Microsurgical treatment, especially in cases of recurrent hemorrhage, remains the treatment of choice.

Case Report

A 24-year-old woman presented with one-week left hemiparesis and left hemihypesthesia, unilateral right ptosis with limited upward gaze, and fixed dilated pupil pointed down and out. She complained of binocular diplopia that was accentuated in upward

terminal gaze and numbness in the left side of the face. There was also persistence of mild flattening of the right half of the face, with the loss of forehead wrinkles and horizontal lines on the same side.

On admission, her cognitive functions were normal. Further examination confirmed left hemihypesthesia, left hemiparesis: left arm 2/5 and left leg 3/5. Reflex response was hyperactive on the left arm (grade 3 on Wexler's scale).

There was dissociated palsy of the right oculomotor nerve with lost ipsilateral direct pupillary light reflex, dissociated palsy of the left trigeminal nerve with preserved pain and temperature sensations, and mild right peripheral facial nerve palsy (grade II on House-Brackmann scale). Corneal reflex was weak on both sides. Left leg was slower in performance of the heel to shin test.

Radiological findings

Magnetic resonance imaging (MRI) of the brain (T1-, T2-weighted images, FLAIR and DWI images)

Correspondence to: Tomislav Sajko, MD, PhD, Clinical Department of Neurosurgery, Sestre milosrdnice University Hospital Center, Vinogradska c. 29, HR-10000 Zagreb, Croatia
E-mail: neurosajko@gmail.com

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demonstrated a popcorn-like lesion surrounded with hematoma in the right half of the mesencephalon. On T2-weighted images, clear distinction between the cavernoma and the surrounding hemorrhage could be seen. There was marked edema of the right half of the mesencephalon (Fig. 1).

Operative procedure

The patient underwent total intravenous anesthesia. Intraoperative neurophysiologic monitoring was performed throughout the operation. The patient was placed in the park-bench position with the ipsilateral mastoid being the highest point in the operation field. Retrosigmoid suboccipital craniotomy with extension to the subtemporal region was performed with skeletonization of the right sigmoid and transverse sinus.

Upon dural opening, the cerebrospinal fluid was released from the cerebellomedullary cistern in order to relax the right cerebellar hemisphere. After retraction of the right cerebellar hemisphere, the VII/VIII complex and anteroinferior cerebellar artery were vi-

sualized. The arachnoid was dissected showing the fifth nerve and the edge of the tentorium. The fourth nerve was dissected from the tentorium edge and the tentorium was transected in order to visualize the supratentorial portion of the mesencephalon. There was yellowish coloration on the right posterolateral mesencephalic surface. The incision was made over this yellowish coloration and at the depth of 3 mm a hematoma was found and evacuated. At the upper border of the hematoma, a rim of the cavernoma was spotted. A boundary between the cavernoma and the yellowish surrounding parenchyma was established. Following this boundary, the cavernoma was removed in a piecemeal fashion.

Intraoperative neurophysiologic monitoring confirmed stable neurophysiologic condition of the sensory and motor pathways throughout the surgery, with evident gradual improvement from pathologic baseline after removal of the cavernoma.

Histopathologic examination confirmed the diagnosis of cavernous angioma.



Fig. 1. Magnetic resonance of the brain. Axial T2-weighted images. Cavernoma in the right half of the mesencephalon with surrounding hemorrhage. The intraparenchymal hemorrhage was closest to the right posterolateral surface of the mesencephalon.



Fig. 2. Multi-slice computed tomography of the brain. Immediate postoperative axial images showing no signs of mesencephalic hemorrhage.

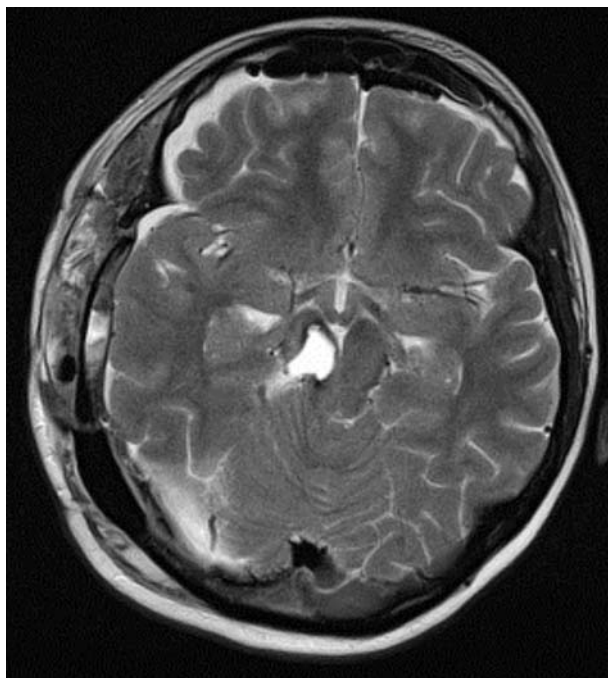


Fig. 3. Magnetic resonance image of the brain 48 hours after the surgery. Axial T2-weighted image demonstrating the cavity after total removal of the mesencephalic cavernoma. There is no sign of the mesencephalon ischemia or edema.



Fig. 4. Magnetic resonance image of the brain. Postoperative axial T2-WI three months after the surgery showing no residual cavernoma, only hemosiderin deposits on the rim of the resection cavity.

Outcome

The immediate postoperative multi-sliced computed tomography (MSCT) showed no signs of residual intraparenchymal hematoma (Fig. 2). Postoperative MRI performed 48 hours after the surgery confirmed total resection of the cavernoma (Fig. 3).

In the first three-month postoperative period, the patient recovered well with complete regression of left hemihypesthesia and total recovery of her left arm and leg strength 5/5.

Reflex response was symmetric and normal on both arms and legs (grade 2 on Wexler's scale). She still had mild right oculomotor palsy. Other cranial nerves were normal. Left leg improved in performance of the heel to shin test. Follow up MRI was performed revealing no signs of cavernoma (Fig. 4).

Six months after the surgery, the patient only complained of episodic gait instability. The right oculomotor palsy recovered completely.

Discussion

Brainstem cavernomas (BSC) account for less than 20% of intracerebral cavernomas and are mostly located in the pons¹⁻³. During the last 4 years, 18 surgical series including a total of 710 patients have been published⁴. Complete excision was performed in 92% of cases. Complication rates were as follows: 42% early neurological morbidity, 84% improvement or unchanged state, and 16% worsening or death at long-term follow up.

The overall hemorrhage rate of all cavernomas is reported to be 2.4% *per patient-year*⁵. Porter *et al.* report on the rate of annual clinical event as high as 10.6% for BSC⁶. A subset of BSC patients may have a relatively benign natural history, while others may rebleed several times in one year^{7,8}. Hauck *et al.* report on a monthly event rate of 8% after second neurological event⁹. Gross *et al.* proposed surgery after the second bleeding event, classifying those rebleeding BSC as aggressive ones⁴. This supports surgical intervention after at least one BSC hemorrhage.

Alternatives are conservative treatment and stereotactic radiosurgery. Conservative treatment is often associated with poor outcome. In a retrospective analysis of 139 patients with BSC during the observation period of up to 25 and 32 years, Fritsch *et al.* found that 20% of the conservatively treated patients died, while there was no lethal outcome in the surgically treated group¹⁰. Stereotactic radiosurgery, as stated by Lunsford *et al.*, is an alternative to observation, but not to microneurosurgery¹¹.

The timing of surgery is debatable. Fahlbusch *et al.* advocate resection of the lesion in the subacute phase¹². Samii *et al.* report no statistical difference in favor of patients who underwent surgery within three months after hemorrhage compared with those in whom resection was performed after three months¹³.

In our case, the decision to operate in the subacute phase was made due to deterioration of the patient's neurological condition. Based on the 2-point method, the lateral supracerebellar infratentorial approach with the patient in the park-bench position was chosen as the most appropriate one¹⁴.

Cavernomas of the dorsal mesencephalon are approached using the supracerebellar infratentorial or occipital transtentorial approach. Sanai *et al.* describe the supracerebellar infra- and supratrochlear trajectory¹⁵. DeOliveira *et al.* describe the medial, paramedial and extreme lateral supracerebellar infratentorial approach¹⁶. The posterolateral portion of the mesencephalon could also be approached *via* subtemporal, transpetrosal or combined trajectory. Whatever the surgical approach, care must be taken not to further damage the surrounding brainstem tissue. Careful hemostasis and inspection for residual lesion is of utmost importance to avoid recurrent bleeding.

Intraoperative neurophysiologic monitoring should be used as the gold standard during BSC operations to avoid nuclear and long tract damages¹⁷. During the surgery, intraoperative neurophysiologic monitoring confirmed stable neurophysiologic condition of the sensory and motor pathways, with evident gradual improvement from pathologic baseline after removal of the cavernoma. The immediate amelioration and almost complete recovery in our patient was, in our opinion, due to cavernoma excision and complete mesencephalic edema regression as clearly seen on early postoperative MRI.

Conclusion

Brainstem cavernomas still present a microsurgical challenge with respectable morbidity. Nevertheless, in cases of recurrent hemorrhage, microsurgical treatment with the goal of complete cavernoma excision remains the treatment of choice.

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Sažetak

POTPUNI NEUROLOŠKI OPORAVAK NAKON NEUROKIRURŠKOG ODSTRANJENJA KAVERNOMA MEZENCEFALONA: PRIKAZ SLUČAJA

K. Rotim, T. Sajko, I. Škoro, M. Zmajević-Schönwald i M. Borić

Kavernozni hemangiomi (kavernomi) su klasificirani kao oblik vaskularnih malformacija središnjeg živčanog sustava. Konzervativno liječenje kavernoma mozgovnog debla je povezano s nepovoljnim ishodom. Operacijsko liječenje kavernoma mozgovnog debla predstavlja izazov zbog pratećeg visokog rizika od neuroloških ispada i značajnog pobola. Prikazuje se slučaj 24-godišnje bolesnice s kavernomom u mezencefalonu kod koje je postignut potpuni neurološki oporavak nakon neurokirurškog odstranjenja kavernoma. Prikazani slučaj potvrđuje da mikroneurokirurško liječenje predstavlja metodu izbora u liječenju bolesnika s jasnim neurološkim ispadom uzrokovanim kavernomima mozgovnog debla koji su opetovano krvarili. Intraoperacijsko neurofiziološko praćenje je neophodno radi maksimalno mogućeg smanjenja oštećenja jezgara i neuronskih putova unutar mozgovnog debla.

Ključne riječi: *Hemangiom, kavernozni, središnjeg živčanog sustava – kirurgija; Mezencefalon; Krvarenje; Oporavak funkcije; Prikazi slučaja*